CONGENITAL HEART DISEASE

Right to left shunt through interatrial septal defects in patients with congenital heart disease: results of interventional closure

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Objective: To study the effects of closure of interatrial communications associated with a right to left shunt in patients with congenital heart disease (CHD) who had a biventricular repair.
Design: Retrospective study.
Setting: Tertiary referral centre.
Patients: 15 patients with CHD with right to left shunt through an interatrial communication: three had repaired tetralogy of Fallot, five had repaired pulmonary atresia with intact ventricular septum, four had Ebstein's disease, and three had other CHDs. Two patients had had a stroke before closure of the interatrial communication.
Interventions: Percutaneous atrial septal defect (n = 6) or persistent foramen ovale (n = 9) closure. All patients underwent an exercise test before and after interatrial communication closure.
Results: Five patients were cyanotic at rest. During exercise, mean (SD) oxygen saturation diminished from 93.9 (3.8)% to 84.3 (4.8)% ($p < 0.05$). Interatrial communication closure led to an immediate increase of oxygen saturation from 93.9 (3.8)% to 98.6 (1.6)% ($p < 0.05$). At a median follow up of three years (range 0.5–5) all but one patient with a residual atrial septal defect had normal oxygen saturation at rest and during exercise. Maximum workload increased from 7.2 (1.9) to 9.0 (2.2) metabolic equivalents ($p < 0.001$).
Conclusions: Percutaneous closure of interatrial communications associated with a right to left shunt allows restoration of normal oxygen saturation at rest, avoidance of desaturation during exercise, and improvement of exercise performance in patients with CHD.

Interatrial communications are usually closed to prevent late right heart failure, arrhythmias, and pulmonary hypertension when they induce right ventricular overload by means of left to right shunt.[1]

Right to left shunt through a persistent foramen ovale (PFO) or an atrial septal defect (ASD) has been reported in several conditions such as the postoperative respiratory distress syndrome, cardiac tamponade, obstructive sleep apnoea, the platypnoea-orthodeoxia syndrome, and right ventricular cardiomyopathy.[2–6] Closure of interatrial communications in these conditions usually improves or resolves symptoms.[5] Although several authors highlighted the importance of suppressing a surgically created right to left shunt in patients who have had a Fontan procedure, no specific reports exist on closure of interatrial communications associated with right to left shunt in children with congenital heart disease (CHD) who have had a biventricular repair.[7]

Right to left atrial shunt occurs when right atrial pressure is higher than left atrial pressure. This can be observed in patients with pulmonary hypertension, right ventricular dysfunction, or diminished right ventricular compliance.[2–6] Right ventricular compliance can be unmasked during exercise. Right to left shunt through an interatrial communication heightens the risk of stroke. In addition, systemic desaturation at rest or during exercise can limit physical ability and impair the quality of life of patients with repaired CHD.[8]

The objective of our study was to present the results of interventional suppression of right to left shunt in 15 consecutive patients with CHD.

PATIENTS AND METHODS

Study population

From January 2000 to December 2004, 15 patients with CHD (median age 13 years, median weight 38 kg) and a right to left shunt through an interatrial communication underwent percutaneous ostium secundum-type ASD (n = 6) or PFO (n = 9) closure. Table 1 lists their clinical characteristics. Patient 14 had a surgically created ASD due to the presence of diminutive pulmonary arteries; this patient was included in the study even though, at time of ASD closure, he had normal oxygen saturation at rest and a minimal decrease of oxygen saturation during exercise. Three patients had repaired tetralogy of Fallot, five had repaired pulmonary atresia with intact ventricular septum, four had unrepaired or palliated Ebstein's disease, and three had other repaired CHD (table 1). Three patients with unrepaired Ebstein's disease had a minor form of the disease, without major incompetence or malformation of the tricuspid valve. Surgical repair of the tricuspid valve was not considered to be indicated in this subset of patients. Only one patient with Ebstein's disease had had a partial cavopulmonary connection because tricuspid valvoplasty was not considered to be useful. Complete repair was preceded by a Blalock-Taussig shunt in patients 5, 10, and 13. Patients 5, 9, and 12 underwent percutaneous and patient 7 underwent surgical Blalock-Taussig embolisation. Patient 9 had Blalock-Taussig embolisation during ASD closure. Patients 1 and 6 had had spontaneous closure of the Blalock Taussig shunt. Patient 8

Abbreviations: ASD, atrial septal defect; CHD, congenital heart disease; METs, metabolic equivalents; PFO, persistent foramen ovale

underwent complete repair of tetralogy of Fallot at the age of 1 month, prosthetic substitution of the pulmonary valve (Hancock no 12) at 6 months, and substitution of the pulmonary outflow at 5 years (homograft 16 mm). She had repeated percutaneous dilatations of the pulmonary outflow and pulmonary arteries and finally she had three stent implantations in the pulmonary conduit and in both pulmonary arteries.

Patients 2 and 15 had had a stroke before ASD or PFO closure (table 1). Patient 2 had had repetitive visual troubles, migraine, and vertigo since the age of 12 years; at the age of 15 years she had a right cerebellar embolism, after which the PFO was closed. She had gradual complete resolution of symptoms.

Patient 15 had had several episodes of pulsatile migraine since the age of 13 years. At 16 years he had a right temporal embolism accompanied by left facial hemiparesis and cognitive troubles. He still has mild sequelae. Most patients became fatigued during mild exercise. Patient 4 had migraine but neurological evaluation and imaging were normal. No patient had a packed cell volume $> 0.48\%$.

Atrial communication closure was considered in the presence of mild or moderate desaturation at rest ($\geqslant 85\%$) accompanied by low flow velocity through the interatrial communication at transthoracic echocardiography or when desaturation occurred during exercise. Patients with increased pulmonary arterial pressure and patients with desaturation at rest lower than 85% were excluded.

Study methods

All but two patients underwent exercise testing at a maximum interval of three months before and after percutaneous closure of the interatrial communication.

Patient 6 did not have an exercise test because of young age and patient 13 was unable to perform it because of severe scoliosis. The exercise test was performed on a stationary bicycle following the Bruce protocol.[9] Maximum workload, maximum heart rate, maximum systolic arterial pressure, and oxygen saturation at rest and at maximal exercise were measured. Maximum workload was expressed in metabolic equivalents (METs) calculated by the formula [(power (W) $\times$ 10.3) + (weight (kg) $\times$ 3.5)]/(weight (kg) $\times$ 3.5). Cyanosis was defined as peripheral oxygen saturation $< 95\%$.

Interatrial communications were closed under general anaesthesia, with fluoroscopic and transoesophageal echocardiographic monitoring. Antibiotic prophylaxis was administered.

Different devices were used according to the anatomy of the interatrial communication. Briefly, through the right femoral vein, a multipurpose catheter and then a guidewire were advanced through the interatrial communication into the left superior pulmonary vein. The stretched diameter of the interatrial communication was measured by inflating a non-compliant balloon over the guidewire through the atrial septum until the interatrial shunt was suppressed.[10] Balloon waist, measured at fluoroscopy and echocardiography, allowed the appropriate device diameter to be chosen. The technique did not differ from that used for closure of interatrial communications with left to right shunt; however, in patients with right to left shunt at rest, closure of interatrial communication was preceded by a test occlusion to exclude a possible significant increment of mean right atrial pressure. A long sheath was then advanced over the wire to the left atrium, the guidewire was retrieved, and the device was inserted into the sheath, advanced, and deployed in accordance with the techniques recommended for each type of device.[11–13] In patients receiving an Amplatzer septal occluder, the device was not upsized as compared with patients with a left to right shunt. When echocardiography confirmed correct device positioning, the device was delivered and a final echocardiogram was recorded to evaluate any residual shunt, to highlight any possible impinging of parts of the device on to cardiac structures, and to rule out the presence of pericardial effusion. A residual shunt was considered to be present if colour Doppler flow mapping showed a left to right or right to left shunt across the interatrial septum. It was defined as trivial (< 1 mm colour jet width), small (1–2 mm colour jet width), moderate (2–4 mm colour jet width), or large (> 4 mm colour jet width).[1]

Age and weight are reported as median and a range; continuous variables are presented as the mean (SD). Individual parameters before and after interatrial communication closure were compared by the two tailed paired *t* test. A two sided $p \leqslant 0.05$ was considered to indicate significance.

RESULTS

Five patients, three with Ebstein's disease (patients 1, 4, and 5) and two with repaired pulmonary atresia with intact ventricular septum (patient 6 and 7), had desaturation at rest. In our series, packed cell volume did not correlate with

Table 1 Characteristics of the patients at cardiac catheterisation

No	Diagnosis	Treatment	Age (years)	Weight (kg)	ASD/PFO	Symptom	O_2 saturation (%)		Packed cell volume
							Rest	Maximal exercise	
1	Pulmonary atresia-IS	BT + valvectomy	11	30	PFO	Fatigue	95	85	0.47
2	Ebstein's disease	None	14.5	51	PFO	Stroke	94	88	0.43
3	Ebstein's disease	None	8	28	ASD	Fatigue	86	80	0.44
4	Ebstein's disease	PCPC	12	38	PFO	Migraine	85	76	0.40
5	Pulmonary atresia-IS	RVOT reconstruction	14.7	44	PFO	Fatigue	96	83	0.42
6	Pulmonary atresia-IS	Interventional, then BT	4	15	ASD	Fatigue	85	ND	0.36
7	Pulmonary atresia-IS	BT + valvectomy	14	40	PFO	None	92	87	0.48
8	Tetralogy of Fallot	Complete repair	12	34	ASD	Fatigue	98	77	0.38
9	Pulmonary valve stenosis	Interventional, then BT	9	30	ASD	Fatigue	96	78	0.39
10	cTGA, VSD, PA	Complete repair	13	45	ASD	None	98	85	0.46
11	Ebstein's disease	None	29	70	PFO	None	98	88	0.48
12	Pulmonary atresia-IS	BT + valvectomy	14	47	PFO	None	96	83	0.48
13	Tetralogy of Fallot	Complete repair	15	44	PFO	Fatigue	95	ND	0.47
14	Tetralogy of Fallot	Complete repair	16	56	ASD*	Fatigue	98	96	0.40
15	Pulmonary valve stenosis	Interventional	16	63	PFO	Stroke	97	94	0.43

*Surgically created.
ASD, atrial septal defect; BT, Blalock-Taussig; cTGA, corrected transposition of the great arteries; IS, intact septum; ND, not done; PA, pulmonary atresia; PCPC, partial cavopulmonary connection; PFO, persistent foramen ovale; RVOT, right ventricular outflow tract; VSD, ventricular septal defect.

Table 2 Haemodynamic data before and after interatrial communication closure

			Before atrial communication closure			After atrial communication closure			
No	Type of device	Device diameter (mm)	Basal O_2 saturation (%)	Basal mean RAP (mm Hg)	Basal RVEDP (mm Hg)	Final O_2 saturation (%)	Final mean RAP (mm Hg)	Final RVEDP (mm Hg)	Residual shunt
1	STARFlex	23	94	6	8	98	7	10	None
2	Amplatzer PFO	25	95	6	8	100	6	10	Small
3	STARFlex	23	86	7	9	96	7	9	None
4	Amplatzer PFO	25	85	8	10	94	8	11	Moderate
5	Amplatzer PFO	35	96	9	12	100	9	12	None
6	ASO	15	84	10	12	100	9	12	None
7	ASO	16	92	9	11	98	10	12	None
8	ASO	14	98	12	14	98	13	15	None
9	ASO	16	96	8	10	98	10	12	None
10	ASO	10	98	6	8	100	6	10	None
11	HELEX	35	98	10	12	100	12	12	Small
12	HELEX	25	96	10	12	100	10	12	None
13	ASO	6	95	12	13	97	12	14	None
14	ASO	24	98	6	8	100	6	8	Small
15	PFO-Star	20	97	6	9	100	5	8	None

ASO, Amplatzer septal occluder; RAP, right atrial pressure; RVEDP, right ventricular end diastolic pressure.

oxygen saturation at rest, probably because desaturation at rest was often mild and present in only five patients. Maximum workload was 7.2 (1.9) METs, maximum systemic arterial pressure was 137 (15.2) mm Hg, and maximum heart rate was 173 (16.8) beats/min. Before ASD or PFO closure eight patients performed a submaximal test. At maximal exercise mean oxygen saturation diminished from 93.9 (3.8)% to 84.3 (4.8)% ($p < 0.05$).

Closure of the interatrial communication was possible in all patients (table 2). It prompted an immediate increase of mean oxygen saturation from 93.9 (3.8)% to 98.6 (1.6)% ($p < 0.05$). Mean right atrial pressure did not change significantly, from 8.3 (1.8) mm Hg to 8.7 (2.1) mm Hg, nor did end diastolic right ventricular pressure. Three patients had a small and one had a moderate residual shunt. No complications occurred.

Exercise test, performed 2.3 (0.9) months after interatrial communication closure, showed that maximum workload increased from 7.17 (1.93) to 9.03 (2.2) METs ($p < 0.001$), whereas maximum systolic arterial pressure (155 (31.8) mm Hg) and maximum heart rate (183 (8.9) beats/min) did not change significantly. Oxygen saturation at maximal exercise increased from 84.3 (4.8)% to 97.5 (2.8)% ($p < 0.001$).

At a median follow up of three years (range 0.5–5 years) all patients were asymptomatic (table 3). There was no recurrence of cerebral events. Migraine regressed in patient 2 and 15. Patient 11 had radiofrequency ablation of an accessory pathway (Wolff-Parkinson-White syndrome) responsible for episodes of supraventricular tachycardia.

All patients with an immediate postprocedural small residual shunt had no residual shunt at follow up transthoracic echocardiography. Patient 4 has a residual ASD with a trivial shunt at rest and mild right to left shunt during exercise (maximum oxygen saturation decreased from 96% to 82%). All the remaining patients have normal oxygen saturation at rest and during exercise.

DISCUSSION

Spontaneous or induced right to left shunt through an interatrial communication is associated with two main possible complications: systemic embolism and systemic desaturation. In adults, several conditions exist in which closure of an interatrial communication has been advocated to prevent or treat symptoms.

Percutaneous closure of PFO is considered at least equivalent to medical treatment in patients with cryptogenic stroke and repeated cerebral embolism.[14] Patients affected by the platypnoea-orthodeoxia syndrome generally completely recover after surgical or percutaneous suppression of the interatrial shunt.[5 15] Other authors reported the results of percutaneous suppression of right to left shunt in patients

Table 3 Ergometric data before and after interatrial communication closure and outcome of patients

	Before atrial communication closure			After atrial communication closure					
No	Basal O_2 saturation (%)	O_2 saturation at maximal exercise (%)	Workload (METs)	Basal O_2 saturation (%)	O_2 saturation at maximal exercise (%)	Workload (METs)	Follow up (years)	Outcome	Events
1	94	85	12.8	98	98	15.7	7	Well	None
2	95	88	4.9	100	98	5.9	6	Well	None
3	86	80	7.3	96	96	10.5	5	Well	None
4	85	76	4.7	94	82	5.6	5	Well	None
5	96	83	5	100	100	7	5	Well	None
6	84	ND	ND	100	ND	ND	4	Well	None
7	92	87	7.6	98	98	9.8	4	Well	None
8	98	77	6.2	98	98	8.8	4	Well	None
9	96	78	9.8	98	98	9.8	3	Well	None
10	98	85	4.9	100	100	7.6	4	Well	None
11	98	88	4.8	100	100	6	3	Well	RF ablation
12	96	83	8.5	100	100	13.5	3	Well	None
13	95	ND	ND	97	ND	ND	2	Well	None
14	98	96	7.3	100	100	7.8	1	Well	None
15	97	94	9.4	100	100	9.4	1	Well	None

METs, metabolic equivalents; ND, not done; RF, radiofrequency ablation of accessory pathway.

with atrial right to left shunt and normal pulmonary arterial pressure.[16] Bassi *et al*[17] reported the successful percutaneous suppression of a right to left interatrial shunt in the setting of a right ventricular infarction. Ebeid *et al*[18] reported the feasibility of percutaneous ASD closure in two patients with pulmonary atresia and intact ventricular septum having a prominent eustachian valve. To our knowledge, however, no reports exist concerning mid term results of percutaneous suppression of right to left shunt in patients with CHD who have had a biventricular repair. In patients with repaired CHD, a right to left atrial shunt through an ASD or a PFO can occur at rest or during exercise, when one or more of the following conditions are present: diminished right ventricular compliance, right ventricular dysfunction, or functional or anatomical tricuspid valve restriction such as in Ebstein's anomaly.

Desaturation at rest is rare in repaired CHD. It can, however, be a prominent physical finding in patients with native or palliated Ebstein's anomaly of the tricuspid valve. The right to left shunt in this setting is the consequence of a number of factors, including interference of right ventricular filling by the redundant valve leaflets and insufficiency of the tricuspid valve into the atrialised portion of the right ventricle.[16] We included in our series three patients with a minor form of Ebstein's disease, whose only symptom was systemic desaturation. None had significant tricuspid valve regurgitation that could have led, after ASD closure, to further enlargement of the right atrium. Indeed, in these patients the risk of repeated stroke was considered important and the closure of the interatrial communication was not thought to prevent a possible future surgical option. In our series, apart from three patients with Ebstein's disease, two more patients with repaired pulmonary atresia and intact ventricular septum had desaturation at rest, probably because of diminished right ventricular compliance and volume.

Cyanosis at rest is typically associated with polycythaemia due to an erythropoietin mediated response to the hypoxic environment. Patients with cyanotic heart disease invariably have reduced exercise tolerance and are at risk of cerebral embolism due to increased blood viscosity and clot formation.[19] Thus, cerebral events in polycythaemic patients should be expected, mostly in the presence of a right to left shunt through the atrial septum. Increased cyanosis or the appearance of cyanosis during exercise is caused by diminished ventricular filling during more rapid heart rates. Patients who are still cyanotic in the absence of pulmonary vascular disease usually are able to exercise only at low intensity levels secondary either to increasing cyanosis with exercise or to pulmonary limitations of exercise.[8]

In our series, right to left interatrial shunting was responsible for cyanosis either at rest or during exercise. In patients with normal oxygen saturation at rest, unmasked cyanosis during exercise showed impaired right ventricular compliance when cardiac output and right ventricular end diastolic volume increased. This series included one patient with mild Ebstein's disease and nine patients with repaired obstructive disease of the right heart. Thus, desaturation at rest can persist in some patients with repaired obstructive disease of the right heart, although desaturation during exercise is more common in this group of patients.

Patients with evident cyanosis at rest and an increase in right atrial pressure higher than 5 mm Hg were not considered to benefit from interatrial communication closure. Excessive increase of right atrial pressure can in fact be detrimental and prompt an increase in central venous pressure, with subsequent development of hepatomegaly and oedema. Unfortunately, at the time of the study, MRI, which would have helped us to evaluate right ventricular size and function, was not available for paediatric patients.

Cerebral events occurred in two patients with mild desaturation either at rest or during exercise. In both, stroke was preceded by migraine, visual troubles, or both. Thus, mild desaturation during exercise should not be underestimated in patients with CHD and the risk of paradoxical embolism should be kept in mind.

Although we did not measure oxygen consumption, we showed that, in our series, work capacity improved early after interatrial communication closure. In addition, easy fatigability regressed in all but one patient with a residual shunt.

It is difficult to study right ventricular function in systole and diastole and, therefore, to evaluate right ventricular compliance with standard echocardiographic methods. Standard exercise testing is, however, a widely used technique that can identify patients who can benefit from closure of an interatrial communication. Indeed, when desaturation occurs only during exercise, symptoms can be scanty. However, closure of the interatrial communication should be taken into consideration in these patients because of the risk of stroke. Standard exercise testing should be recommended for patients with repaired obstructive disease of the right heart and in those with minor forms of Ebstein's disease. However, in patients with Ebstein's disease suppression of a right to left shunt can potentially lead to further enlargement of the right atrium and development of arrhythmias. Long term studies can probably clarify this issue.

In conclusion, our data show that percutaneous closure of interatrial communications associated with a right to left shunt allows restoration of normal oxygen saturation at rest, avoidance of desaturation during exercise, and improvement of work capacity. Evaluation of oxygen consumption and more sophisticated studies of right ventricular compliance should be encouraged.

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IMAGES IN CARDIOLOGY

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Infective pseudoaneurysm of a ruptured sinus of Valsalva as an unusual cause of myocardial infarction by compression of the right coronary artery

A 25 year old man was referred to our institution for chest pain associated with fever despite oral antibiotic treatment. ECG showed evidence of a semi-recent inferior myocardial infarction. Transthoracic and transoesophageal echocardiography demonstrated a circumferential pericardial effusion, severe hypokinesia of the left ventricular inferior wall, and a mobile mass on a bicuspid aortic valve associated with grade 2 aortic regurgitation. A large cavity (5 cm × 3 cm) was noted next to the right coronary cusp with a fistulous track from the ascending aorta. Multislice spiral computed tomography (16-slice Lightspeed, General Electric) demonstrated a thoracic aortic pseudoaneurysm with a 1 cm-wide neck from a ruptured right sinus of Valsalva causing external compression of the right coronary artery (upper and lower panels: An, aneurysm; Ao, aorta; LAD, left anterior descending coronary artery; RCA, right coronary artery; MI, myocardial infarction; PE, pericardial effusion). Urgent surgery confirmed compression of the right coronary artery by a large mycotic pseudoaneurysm related to a ruptured right sinus of Valsalva and aortic infective endocarditis. The aortic valve was replaced by a mechanical valve with closure of the pseudoaneurysm by two pericardial patches.

In this report, myocardial infarction was related to an elongation of the right coronary artery by a large mycotic pseudoaneurysm of a ruptured sinus of Valsalva. Very few cases of coronary artery compression associated with aortic pseudoaneurysm have been reported so far. Physicians have to be aware of this unusual complication requiring urgent surgery. Multislice spiral computed tomography may have an important role in achieving an accurate and rapid diagnosis, and guiding surgery.

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